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Case Report

Primitive neuroectodermal kidney tumour

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Primary neuroectodermal tumours of the kidney are very rare. We report the sixth case of such a tumour in an adult male.

CASE REPORT:

A 39 year old man presented with frank haematuria and right loin pain. Clinical examination and serum creatinine were normal. There was no excretion from the right kidney on intravenous urography. Ultrasound and CT scan revealed the presence of a 10 cm diameter solid lesion in the upper pole of the right kidney. Two lesions, suggestive of metastasis, were noted in the right lung. Isotope bone scan was normal.

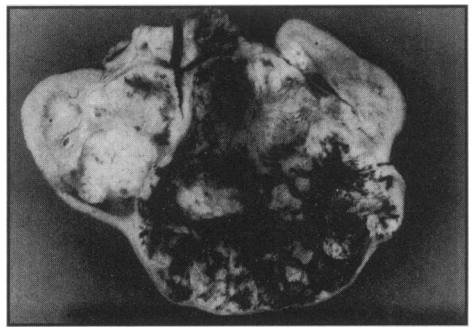

Fig. 1 Right kidney on cut section showing solid tumour with areas of haemorrhage and cystic degeneration. Renal vein is occluded with tumour.

A right radical nephrectomy was performed. The tumour was mostly solid with areas of haemorrhage and cystic degeneration (Fig 1). It had infiltrated the renal capsule and also had invaded and completely occluded the renal vein. On microscopy it was composed of islands and sheets of small pleomorphic cells most of which were ovoid with scant cytoplasm, variable nuclei but lacking prominent nucleoli. There was significant mitotic activity, and both necrosis and apotosis were seen. Vascular invasion was quite prominent. Immmunostains for vimentin were negative but immunostains for HBA-71 stain (MIC 2 gene product) was sufficiently positive on tumour cell cytoplasmic membrane to confirm the diagnosis of primary primitive neuroectodermal tumour of the kidney.

The patient has been treated with eight courses of combination systemic chemotherapy (vincristine, adriamycin and cyclophosphamide) postoperatively. Radiotherapy has been reserved for any local recurrence. Follow up computerised

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tomography showed complete resolution of the pulmonary lesions and no evidence of local recurrence twenty months after operation.

DISCUSSION

Primitive neuroectodermal tumours are a group of neoplasms which are presumed to arise from pluripotential neural crest cells; they present predominantly in childhood and young adults.[1] These tumours are known to originate in the central nervous system or peripherally in the adrenal glands and sympathetic ganglia. Besides these common sites there are sporadic reports of such tumours arising from peripheral nerves, musculoskeletal system, skin and urogenital system including kidneys.[2] Histologically they are composed of small round cells with focal Homer-Wright rosette formation, and on electron microscopy show interdigitating cytoplasmic processes, a variable number of neurosecretary granules and microtubules.[1] Immunohistochemistry plays a significant role in establishing the diagnosis by demonstrating the expression of various neurone specific markers. These include synapsin I, synaptophysin, neural cell adhesion molecules (N-CAMs), neuron-specific enolase (NSE), nestin, vimentin, neurofibrillary protein (NF), glial fibrillary acid protein (GFAP) and less commonly protein S-100.[3, 4, 5] A variety of numerical and structural abnormalities of chromosomes have been identified including a reciprocal translocation between chromosome 11 and 12 (11; 22) (q24; q12) and the presence of an isochromosome seventeen and trisomy of chromosome 1q.[6]

Peripheral primitive neuroectodermal tumours are highly aggressive neoplasms which not only have a tendency to recur locally after excision but also have a predilection to metastasize to distant sites like liver, lungs, pleura, peritoneum, bone and lymph nodes.[7] Primary neuroepithelial tumours of the kidney are rare, and only five cases of adult primitive neuroectodermal tumours of the kidney have been reported. They are more common in males. Positive immunostaining by HBA-71 distinguishes these tumours from blastemal tumours with which they can be confused histologically. There is no standard treatment regimen for such tumours. Two of the reported patients with advanced local disease at presentation died within a year in spite of multimodal treatment. One patient with disease localized to the kidney was reported alive five years after surgery and postoperative radiotherapy.[8] Our patient has had a complete response to systemic chemotherapy and is currently disease free 20 months after surgery.

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ERRATUM

Intravenous Mercury: a three year follow up:

W J A Anderson: Ulster Medical Journal 1993; 63; 180-183.

It has been brought to our attention that the case referred to in this report had been diagnosed and treated in a different hospital in Northern Ireland to the one stated in the case report. Clinical biochemical advice and the management was provided by Dr Pooler Archbold.

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